

The scaphocephalic skull of an adult male

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The paper presents abnormal craniofacial morphology of an adult male afflicted with premature closure of the sagittal suture. The skull is well preserved and there are no visible traits of surgical management which would be aimed to correct cranial deformation. In consequence of the restricted cranial development, some diameters of the skull were significantly altered. Basically, cranial vault morphology fits apparently to the scaphocephaly, whereas the basicranium and viscerocranium are altered only in minor degree. (Folia Morphol 2014; 73, 1: 92–98)

Key words: craniosynostosis, sagittal synostosis, sagittal suture

INTRODUCTION

The term 'scaphocephaly' refers to the condition where the head is disproportionately long and narrow. It is caused by the premature closure of the sagittal suture, which normally remains patent till the 3rd or even 4th decade of life. Premature closure of a cranial suture restricts cranial growth in perpendicular direction towards obliterated suture. However, compensatory growth occurs at neighboring unfused sutures, and the skull enlarges disproportionately [11]. In the case of sagittal synostosis that occurs between 2 parietal bones, transverse expansion of the cranium is considerably limited. In consequence, it gives a keel-like shape of the skull which may have midline bony ridge over the interparietal suture region, biparietal and bitemporal narrowing and occipital prominence [19, 21].

In the contemporary population, craniosynostosis occurs approximately once per 2000 life births [23]. Scaphocephaly is the most common form of isolated craniosynostosis, accounting for 40–60% of single suture synostosis. The incidence of sagittal synostosis in the population is approximately 1 in every 4200 births, with a male to female ratio of 3:1 [16].

Infants born with the sagittal synostosis usually need surgical treatment in order to release the

involved suture and reshape the malformed bones of the skull. Surgical correction of craniosynostosis is usually performed between 4th and 8th month of age. Otherwise, altered shape of the skull retains throughout the whole life, and may be a reason of neurologic problems [23].

Nowadays, adult cases of craniosynostosis are rarely observed in population because majority of children with that pathology are subjected to surgical management. There is only limited number of reports on adult craniosynostosis [14, 17]. Therefore we present a case of an adult unoperated scaphocephalic skull and compare its diameters to the normal skulls and similar cases described in the literature. In the current study we focused on analysing the shape and size of the brain case, which morphology results from the sagittal synostosis.

MATERIALS AND METHODS

The scaphocephalic skull of the adult male individual derives from the craniological collection housed in the Anatomical Museum of the Department of Anatomy, Collegium Medicum of the Jagiellonian University in Cracow. Unfortunately, the origin of the skull and its chronology are unknown. The investigated skull is complete and well preserved. Sexing was

Table 1. Measurements performed on scaphocephalic skull

	Measurement	Description
1.	Maximum cranial length (g-op)	Direct distance measured between glabella (the most forward projecting point of the forehead in the midline at the level of the supraorbital ridges) and opisthocranium (the point in the midline of the cranium that projects farthest backward)
2.	Maximum cranial breadth (eu-eu)	Maximum width of skull perpendicular to midsagittal plane wherever it is located, with the exception of the inferior temporal lines and the area immediately surrounding them
3.	Bizygomatic diameter (zy-zy)	Maximum breadth across the zygomatic arches, perpendicular to the median plane
4.	Midfacial width (zm-zm)	The shortest distance between right and left zygomaticomaxillare (located externally at the lowest extent of the zygomaticomaxillary suture)
5.	Basion-bregma height (ba-b)	Direct distance from the lowest point on the anterior margin of foramen magnum to bregma (the junction of the sagittal and coronal sutures at the top of the skull)
6.	Upper facial height (n-pr)	Direct distance from nasion (the intersection of the fronto-nasal suture and the median plane) to prastion (the most anteriorly prominent point, in the midline, on the alveolar border, above the septum between the central incisors)
7.	Face height (n-gn)	Direct distance from nasion to gnathion (the lowest point in the inferior border of the mandible at the median plane)
8.	Minimum frontal breadth (ft-ft)	Direct distance between 2 frontotemporale (located at the most anterior point of the temporal line on the frontal bone)
9.	Frontal chord (n-b)	Direct distance from nasion to bregma taken in the midsagittal plane
10.	Parietal chord (b-l)	Direct distance from bregma to lambda taken in the midsagittal plane
11.	Occipital chord (l-o)	Direct distance from lambda (the junction of the sagittal and lamboid sutures) to opisthion (the middle of the posterior, or dorsal, margin of the foramen magnum)
12.	Biasterionic width (ast-ast)	The shortest distance between right and left asterion (the common meeting point of the temporal, parietal, and occipital bones)
13.	Arc length (n-b)	Arch length measured along the squama of the frontal bone between nasion and bregma in the midsagittal plane
14.	Arc length (b-l)	Arch length measured between bregma and lambda along the parietal bone in the mid-sagittal plane
15.	Arc length (l-o)	Arch length measured between lambda and opisthion along the squama of the occipital bone in the mid-sagittal plane

determined by evaluation of the morphology of the bones of the skull. Marked muscular attachments to the bones, strongly developed temporal lines, prominent superciliary ridges, well-developed external occipital protuberance and the shape of mandible express male character of the investigated skull.

The bones of neurocranium and splanchnocranium are heavily mineralised and show no evidence of any perforations. There are no signs of cranial suture fusion with the exception of the sagittal suture. The non-obliterated sutures, ossified sphenoccipital cartilage, and worn teeth suggest that this individual might have died approximately at the age of 25–30 years.

Basic anthropological measurements were taken with an appropriate instrument (sliding caliper, spreading caliper and flexible tape), respectively to the methodology [18]. Because of the absence of the sagittal suture, the bregma and the lambda landmarks

were accepted arbitrarily as the point of intersection of the coronal and lambdoid sutures with the median sagittal plane. Craniofacial measurements performed on the scaphocephalic skull are presented in Table 1.

The proportions between cranial diameters were expressed by 3 cranial indices: breadth-length index ($eu-eu/g-op \times 100$), height-breadth index ($ba-b/eu-eu \times 100$) and height-length index ($ba-b/g-op \times 100$). The ratio of the cranial breadth to the cranial length was used as the standard descriptor of the skull elongation.

All anthropological measurements performed on the investigated skull were compared to the normatives of the adult male dolichocephalic skulls regarded as the reference group [27].

RESULTS

The most prominent feature of the investigated skull is extremely long and narrow brain case ended

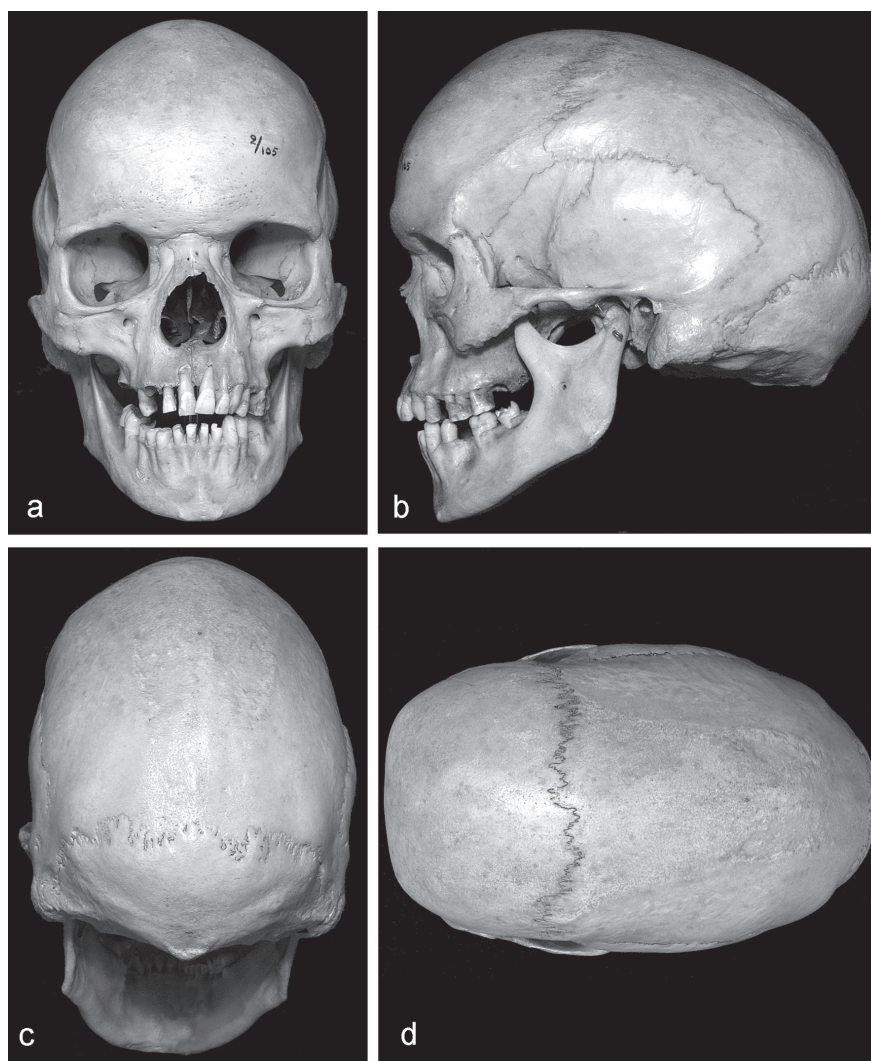


Figure. 1. A scaphocephalic skull of the male, adult individual; **A.** Anterior view; **B.** Lateral view (left side); **C.** Posterior view; **D.** Superior view.

with a bulged occiput. Lateral view of the skull presents typical boat shape of the cranium. Viewed from above and posteriorly the skull is narrow, and shows completely fused sagittal suture and non-obliterated coronal and lambdoid sutures. In turn, frontal view of the analysed skull presents high forehead, long and narrow face, relatively big orbits and wide piriform aperture (Fig. 1).

Measurements of the investigated scaphocephalic skull compared with normal dolichocephalic skulls of the male adults (reference group) are presented in Table 2.

The maximum cranial length ($g-op = 201$ mm) exceeds considerably normative value ($g-op = 188.9$ mm), and also the maximum cranial breadth is decreased ($eu-eu = 127$ mm) comparing to normal doli-

chocephalic skulls ($eu-eu = 136.9$ mm). However, the biggest contrast between measurements of the scaphocephalic skull and normal dolichocephalic skulls is related to the size of the parietal bones. Both the parietal chord ($b-l$) and the parietal arch (arc $b-l$) revealed the biggest differences towards the normal skulls: 26.2 mm and 28.6 mm, respectively (Table 2). The frontal chord ($n-b$) of the investigated scaphocephalic skull is only 5 mm longer than at the reference skulls (120 mm vs. 115 mm, respectively), but the arc length of the frontal bone (arc $n-b$) is 15.2 mm bigger (147 mm vs. 131.8 mm, respectively).

In the studied skull also significant metrical changes occurred within the occipital bone. On the contrary to the frontal bone, both the arc length (arc $l-o$) and the occipital chord ($l-o$) are smaller than normative

Table 2. Craniofacial measurements (in millimetres) and indices of the scaphocephalic skull compared with normal dolichocephalic skulls of the male adults (reference group)

Measurement and index	Scaphocephaly x	Reference group			Contrast x — ref. mean
		Mean	SD	n	
g-op	201	188.9	4.79	30	+12.1
ba-b	130	139.2	6.69	28	−9.2
eu-eu	127	136.9	3.73	30	−9.9
ft-ft	103	97.0	5.92	30	+6.0
ast-ast	103	110.1	5.10	28	−7.1
n-b	120	115.0	4.63	29	+5.0
b-l	144	117.8	6.33	30	+26.2
l-o	85	97.5	4.90	29	−12.5
arc n-b	147	131.8	6.46	29	+15.2
arc b-l	160	131.4	8.54	30	+28.6
arc l-o	110	120.2	7.16	29	−10.2
n-gn	113	117.8	5.59	10	−4.8
n-pr	60	69.3	5.70	23	−9.3
zy-zy	130	129.8	5.27	19	+0.2
zm-zm	91	96.3	4.83	22	−5.3
eu-eu/g-op × 100	63.2	72.5	1.79	30	−9.3
ba-b/eu-eu × 100	102.4	101.8	4.78	28	+0.6
ba-b/g-op × 100	64.7	73.8	3.03	28	−9.1

values of the reference skulls (Table 2). Moreover, the occipital bone does not manifest as the occipital knob, which is typical effect of the cranial deformity resulted from premature suture obliteration [11].

The abnormal cranial shape and size is not only confirmed by absolute values of the diameters and arches of the frontal, parietal and occipital bone, but also by the cranial indices. The ratio between maximum cranial length and breadth (cephalic index) is significantly lower in the case of scaphocephaly (63.2 vs. 72.5 of normal skulls). Such a value indicates on abnormal elongation of the cranial vault in the antero-posterior direction, with mutual biparietal narrowing. In turn, the ratio between the maximum cranial height and maximum cranial length or breadth indicates on the extremely convex calvaria of the investigated skull.

Elongation of the neurocranium in the sagittal plane is followed by less obvious compensatory changes of the facial measurements. The face height (n-gn) of the investigated skull is smaller only by 4.8 mm comparing to normal skulls (113 mm vs. 117.8 mm of normal skulls). The upper face dimension (n-pr)

shows a bigger difference (60 mm of the examined skull vs. 69.3 mm of reference skulls).

DISCUSSION

Scaphocephaly resulting from premature closure of the sagittal suture is one of the types of cranial deformation documented in osteoarcheological materials from different historical periods [7, 13, 14, 28]. The skull being the subject of the present study is of unknown historical and geographical origin. As a museum specimen, it can only serve for a case anatomical study, but delivers valuable information how the cranial morphology was altered in response to the sagittal synostosis untreated surgically.

The investigated scaphocephalic skull is a vivid example of compensatory growth that occurred mainly in the antero-posterior direction. As a result of the abnormal developmental process, one may observe which parts of the skull underwent critical morphological modification in response to anatomical constraints, caused by cessation of head growth in perpendicular direction to the obliterated suture. Early closure of the sagittal suture, which normally is

responsible for transverse cranial growth, caused that skull expanded in the antero-posterior direction [20, 26]. In the studied case parietal bones are extremely elongated sagittally, what makes the whole skull excessively long. The extensive growth of the parietal bones influenced on the size of articulating bones: the frontal and the occipital. In this case, the coronal suture played a crucial role in the compensatory growth of the cranium, which is manifested in increased length of the frontal arch. Moreover, this view may be supported by well visible local extensions within the edges of the parietal bones, which are directed anteriorly. Probably the antero-posterior orientation of bony spicules within the coronal suture reflects driving forces derived from the growing brain, which caused bulging of the frontal bones. In contrast to the frontal bone, the occipital bone displays the reduction in diameters, therefore the primary site of the compensatory growth was located anteriorly.

According to Weber et al. [28], who measured 18 scaphocephalic skulls, mean maximum cranial length was 203 mm, while mean maximum cranial breadth was 127 mm. These values are very close to the same diameters of the analysed scaphocephalic skull. Cranial length measured in scaphocephalic skull from California dated on the Later Middle Horizon was similar to the analysed specimen (203 mm), but the greatest cranial breadth was narrower (120 mm) [9]. Djurić-Srejić [7] described the biggest scaphocephalic skull of an adult which was found in China. The maximum cranial length of Chinese skull was 230 mm, and maximum cranial breadth was 137 mm. The smallest scaphocephalic skull was found in Germany with maximum cranial length of 177 mm [28].

The specimens of adult skulls with premature sagittal synostosis suggest that compensatory growth of the cranium in antero-posterior direction may exceed normal maximal cranial length more than 20 mm, if we accept that maximum cranial length of the normal dolichocephalic skull is 180 mm. The mean maximum cranial diameter of the normal skulls estimated by Weber et al. [28] as 180 mm in German skulls ($n = 40$) is lower by 23 mm than the mean maximum cranial diameter of scaphocephalic skulls measured by those authors. The Chinese scaphocephalic skull is 53 mm longer than Chinese normal skulls ($n = 81$), for which the mean maximum cranial diameter was 177 mm [7]. In our dolichocephalic skulls mean maximum cranial length was bigger (189 mm), therefore,

the difference between the reference group and the analysed skull is lower — 12 mm.

An abnormal cranial shape in the scaphocephaly is particularly expressed by the ratio between the maximum cranial length and breadth (63.2 vs. 72.5 in the case of normal skulls), and the ratio between the maximum cranial height and length (64.7 vs. 73.8 in the case of normal skulls). Both indices of the scaphocephalic skull are smaller of 9 units comparing to normative values, therefore we regarded that specimen as a distinct morphotype which is out of normal biological variation. A similar trend was described by other authors, but the difference between scaphocephalic skulls and the reference group in the cranial index ranged from 3.1 to 17.1 units [7, 9, 28].

The elongation of a scaphocephalic skull is also manifested by a sagittal suture measurement. The mean length of the sagittal suture in the analysed scaphocephalic skull was longer than normative values (144 mm vs. 117.8 mm, respectively). The difference in the mean length of the sagittal suture, measured in German skulls, is statistically significant (143 mm in scaphocephalic skulls vs. 118 mm in normal skulls) and almost similar to our comparison [28].

We also observed the overgrowth of the frontal bone, which was manifested by increased length of the frontal arch and breadth of the frontal bone (Table 2). In contrary to the frontal bone, the occipital bone does not manifest typical symptoms of scaphocephaly such as an occipital knob or an increase of an occipital chord. Albright et al. [1] described the mechanism of premature fusion of sutures. They indicated that craniosynostosis begins in one area of a suture and the ossification progresses ultimately along the suture. Although the ossification of entire sagittal suture has been observed in the analysed scaphocephalic skull, craniometrical results justify the suggestion that a primary site of the compensatory growth involved mainly frontal and parietal bones.

The dimensions of the face in the studied scaphocephalic skull appear to be almost within the range of normal skulls (zy-zy) or slightly lower (n-gn, n-pr) (Table 2). These results support the views of Kohn et al. [13] and Djurić-Srejić [7], who stated that scaphocephaly does not have significant effect on face dimensions but a trend is evident in reducing relative size of a face. Our craniometrical data suggest that scaphocephalic conditions mostly affected dimensions of the cranial vault. Other researches confirm that statement as well [9, 25].

The studied skull delivers the evidence for cranial deformation, which persists to adolescence. Severe skull deformity indicates the lack of surgical treatment, which could correct abnormal shape of the calvaria. This may suggest that the skull can be dated to the past historical periods. In the past, the possibility of surgical management was extremely limited, therefore operations of the head were hazardous. The first reported operation for correction of craniosynostosis was performed in 1890 by Marie-Lannelongue, but the modern era of the surgical treatment of craniosynostosis began in 1970. Nowadays, craniectomy and bone remodelling became standard procedures administrated in the case of craniosynostosis in order to reshape the skull and alleviate the brain to grow, and are carried out in childhood.

In the case of studied scaphocephaly nothing is known about his origin and life style. Theoretically, that individual could suffer from neurological defects which accompany craniosynostosis. Infants and children with scaphocephaly more frequently have cognitive dysfunction, deficits in speech and language learning, working memory, attention and visual-spatial planning [12, 16]. The other common symptoms related to craniosynostosis include: headache, emesis, visual disturbance, irritability, bulging anterior fontanelle, and altered mental status [6]. Those neurocognitive deficits have been associated with increased intracranial pressure and mental retardation.

Unfortunately, we are unable to verify the abovementioned presumptions, especially that many patients with the isolated sagittal synostosis display normal intellectual function [5]. Referring to the contemporary researches, we assume that studied individual with the sagittal synostosis could display anatomical changes of the brain [3, 10, 12]. Although the brain of individuals with craniosynostosis seems to be 'normal' if we take into account the presence of component structures, is abnormal in shape [2]. Lack of bone growth at the fused sagittal suture can affect brain growth and may lead to subsequent injury to the brain, including both cortical and subcortical structures [12]. Aldridge et al. [3] and colleagues show posterior directed elongation of the brain, in both forebrain and hindbrain structures associated with premature fusion of the sagittal suture. Elongation of the brain and related prominence of the forehead and occiput may cause changes in the dorsolateral frontal region of the brain and the resultant problems with working memory and

planning [24]. Constriction of the growing occipital lobes in scaphocephaly (observed also in studied skull) may narrow the posterior area of the brain, which may lead to a deficit of the visual cortex [8]. Also, a constriction of the brain has been observed between the left Sylvian fissure and the left central sulcus [3]. Abnormalities located in the left temporo-parietal and occipito-parietal cortex, including the areas of Sylvian fissure, are linked with dyslexia. Children with sagittal synostosis resemble children with attention deficit-hyperactivity disorder (ADHD), who usually demonstrate reading comprehension and language problems. Other researches show that scaphocephaly is commonly accompanied by a downward displacement of the cerebellar tonsils through the foramen magnum (Chiari I malformation). Patients with Chiari I malformation suffer from headache, vomiting and somnolence [10, 15, 22]. It had been also noted that patients with sagittal synostosis have increased intracranial volume when compared to a normal population [4].

Moreover, studies of Aldridge et al. [3] suggest that morphologic changes of the brain are not limited to the areas under the fused suture. Neuropsychological deficits seem not to be related to a single anatomic malformation, but rather, are associated with cumulative response to multiple structure changes [12]. It should be also noted, that cognitive functions are associated with neural plasticity, compensatory processes, behavioural and environmental factors, therefore neurobehavioural outcomes of patients with craniosynostosis may not only have relation to the fused suture [12]. Because the neurological defects do not affect all cases of the scaphocephaly, we are inclined to the view that individual, whose skull was a subject of our study, revealed normal mental function.

CONCLUSIONS

In conclusion we would like to stress that incidence of scaphocephalic skulls in prehistorical and historical populations has not been estimated, specially, due to rare occurrence of those cranial deformities in skeletal materials. Also, there is no evidence of a child scaphocephalic skull in archaeological materials described in the literature. Children's skeletons are less ossified, therefore remain poorly to present days. The present case represents a rare finding in paleopathological material and provides new data on cranial and facial morphology in adult untreated scaphocephaly.

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